Desert Hematuria

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HPI

- 24 y.o. M referred from Urology for perisitent gross hematuria x 10 months
- First noticed while stationed in Kuwait (had since be transferred to Germany)
- “Brown” urine getting darker throughout void without dysuria
- Dull right flank pain x 2 months radiating to groin

HPI (cont.)

- No history of trauma or stones
- No fevers, URI, rashes
- No hematemesis, hemoptysis
- Initially referred to Urology 5 months after onset: IVP, cystoscopy, CT abd/pelvis normal except small L simple renal cyst
- sickle prep negative, complements nl

HPI (cont.)

- Repeat cystoscopy showed ? bleeding localized to R ureteral orifice
- PMHx: occasional sinusits
- Meds: occasional use of NSAIDs, NKDA
- Habits: no tobacco, EtOH, illicits
- FHx: no renal dz or deafness, +HTN, uncle with heart value replacement in 20s,
- SHx: active-duty USA Sgt. , married, two children in GH

Physical Examination

- Young fit BM in NAD
- afebrile, BP 125/70
- mildly tender to palpation in right lateral abdominal wall and pelvis
- rest of exam normal

Labs

- Cr = 1.1, uric acid = 4.7
- Hct = 42, MCV = 87, plts=162
- PT/PTT normal
- U/A = tea-colored, SG 1.027, large blood, 2+ protein Ssa 50 mg%
- urine sediment = TNTC RBCs with nl morphology, occasional WBCs, no organisms, no casts

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Initial Workup

- CXR normal
- Urine cx: negative
- 24-hr urine: CrCl = 155 ml/min, Protein = 1.1 gms, Ca 146 mg (100-300 nl), Uric Acid = 694 (250-750 nl)
- Sickledex: negative. HgB EP: AA.
- Ureteroscopy: Randall plaques on papillae of right kidney (? Specificity for stones).

Clinical Course I

- Recommended to increase fluid intake and continue clinical f/u
- Placed on 30-day course of Amicar by Urologist in Germany, reported increasing hematuria, continued flank pain
- Returned in 3 months for incapacitating pain

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Workup II

- Renal arteriogram: normal
- Ultrasound-guided percutaneous renal biopsy: skeletal muscle
- CT-guided percutaneous renal biopsy performed

Biopsy Summary

- 44 glomeruli, 1 sclerotic
- Rare proximal tubule epithelium within Bowman's space and glomerular tuft
- Normal tubules, interstitium, arterioles.
- EM: normal GBM thickness, scattered focal podocyte fusion, no deposits
- IF: trace IgM, C3

Diagnosis:

Essential Glomerular Hematuria

with

Loin Pain Hematuria Syndrome

Clinical Course II

- Returned to Germany, continued to have refractory pain.
- Returned to Nephrology 6 months later for medical separation from Army.
- Started on ACE inhibitor, with possible improvement in pain

Intraglomerular Tubular Epithelial Cells (ITEC)

- Unknown pathogenesis, associated with heterogeneous clinical entities
- Three histological patterns
  - tubular metaplasia
  - epithelial cell “reflux” into Bowman’s space
  - tubular cell “emboli” into capillary loops

ITEC (cont.)

- One series reports incidence in 111/202 (55%) consecutive renal biopsies. 110/111 (99%) had gross or microscopic hematuria. 110/123 (89%) of cases with hematuria had ITEC. (Valdes and Zhang, 1987.)
- No reported specific association with loin pain.
- Unknown prognostic significance.

Loin Pain Hematuria

- First described by Little *et al.* in 1967
- Triad: loin pain, hematuria, low-grade fevers.

Loin Pain Hematuria
Epidemiology

- Incidence unquantified, described as “uncommon” and “rare”.
- > 90% of cases are in women
- Age of onset: mid 20s (range 8-50)

Loin Pain Hematuria
Clinical Features

- Loin pain may precede hematuria by years, and *vice versa*.
- Unilateral flank pain radiating to groin, abdomen, or thigh. Can become bilateral.
- Pain may be incapacitating
- May have low-grade fevers, dysuria, frequency

Loin Pain Hematuria
Clinical Features (cont.)

- Attacks variable in frequency (daily to yearly) and duration (hours to weeks).
- Hematuria: can be gross or microscopic, persistent or intermittent.
- Physical Exam: low-grade fevers, flank tenderness.
- Labs: normal Cr and concentrating ability. Normal electrolyte secretion. Proteinuria < 1.5 gms/day.

Loin Pain Hematuria Associations

- Initial reports of relationship to oral contraceptives, now in question.
- Platelet dysfunction: various reports of abnormalities in platelet and coagulative function.
- Psychiatric associations controversial: has been proposed as a somatoform pain disorder (Lucas *et al.*, 1995).

Loin Pain Hematuria
Diagnosis

- Diagnosis of exclusion
- Renal angiogram: originally small vessel abnormalities described, more recent studies seem to refute this.
- Biopsy: variable, mild/moderate, nonspecific findings. Arteriolar C3 deposition may be most reproducible finding. One case series reports 7/15 pts with thin GBM disease (Hebert *et al*., 1996)

Loin Pain Hematuria
Pathogenesis

- Bottom line: unknown.
- Proposed mechanisms:
  - immune-mediated ischemia
  - thrombosis-mediated ischemia
  - atherosclerosis-mediated ischemia
- Proposed mechanisms for pain:
  - tubular obstruction, backflow, renal swelling

Loin Pain Hematuria
Differential Diagnosis

- Nephrolithiasis/Urolithiasis
- Obstructive uropathy
- Pyelonephritis
- Acute GN
- Vascular: Renal infarction, papillary necrosis, AVM, renal vein thrombosis.
- Mass: Cyst/cyst rupture, neoplasm.
- Trauma

Loin Pain Hematuria
Treatment

- Analgesics
- Splanchnic nerve blockade: short term relief only
- Renal autotransplantation : controversial efficacy (~10% or greater recurrance).
- Cessation of oral contraceptives/HRT.
- Warfarin: benefits unproven
- ACE inhibitors

Loin Pain Hematuria
Prognosis

- No increased risk of renal failure.
- Overall mortality unchanged.
- Morbidity related to variable response to analgesic interventions.
- In one series, 30% had spontaneous remission by 3-4 years (Aber and Higgins, 1982).

References

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